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Benign metastasizing leiomyoma

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CASE REPORT

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Benign metastasizing leiomyoma

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Abstract

Benign metastasizing leiomyoma (BML) is a rare condition, affecting predominantly reproductive-age females with uterine leiomyomata and is most often associated with multiple benign-appearing smooth muscle tumors in lungs. We report herein a case of a 38-year-old woman who presented with multiple uterine fibroids for which hysterectomy was carried out on her. Postoperatively, she developed left-sided pleural effusion. Computed chest tomography (CT) scan revealed multiple nodules in both lungs and pleurae. Histopathology of one of the pleura-based nodules revealed a neoplasm composed of interlacing fascicles of spindle cells with uniform nuclei. The tumor cells were positive for alpha-smooth muscle actin and negative for CD34 immunohistochemical stain.

Keywords: Benign metastasizing leiomyoma, lung nodule, metastasizing leiomyoma

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Introduction

Benign metastasizing leiomyoma (BML) is a rare lesion affecting females with history of uterine leiomyomata and are most often characterized by benign-appearing smooth muscle tumors in the lungs and other distant sites including soft tissue, lymph nodes, mesentery, bones, central nervous system (CNS) etc. [1] It usually occurs in young females in their reproductive age. [2] We report herein a case of synchronous BML in a patient with multiple uterine leiomyomata.

Case Report

A 38-year-old woman presented with a large pelvic mass. Clinical and ultrasonographic examination revealed multiple fibroids. Her hysterectomy and unilateral salpingo-oophorectomy was performed. Gross examination of the hysterectomy specimen revealed a large distorted uterus, which on serial sectioning showed multiple mural and subserosal fibroids. The largest fibroid measured 17.5 × 14.5 × 10 cm in dimension with a tan-gray, firm and whorled cut surface. No necrosis was seen on gross examination. Other fibroids ranged from 1.0 to 7.0 cm in diameter, all with tan-gray, firm and whorled cut surface. Endometrial cavity was distorted; however, no growth was seen in the endometrial cavity. All fibroids were adequately sampled. Histological examination revealed lesions composed of interlacing fascicles of smooth muscle cells with uniform elongated cigar-shaped nuclei. The mitotic count was 1 per 10 HPF. Evidence of coagulative necrosis, focal or diffuse atypia or increased mitotic activity was not seen.

Postoperatively, she developed left-sided pleural effusion. Computed tomography (CT) scan revealed multiple, bilateral lung nodules with a suspicion of metastatic disease. Computed tomography assisted Tru-Cut biopsy of one of the left lung nodules was performed, which revealed scanty non-diagnostic material. Open biopsy of one of the pleural-based nodules was then performed. On gross examination, there was a well-circumscribed nodule measuring 1.8 × 1.5 × 1 cm. Cut surface was tan-gray, firm and whorled in appearance. Histological examination revealed a lesion composed of spindle cells with elongated cigar-shaped uniform nuclei [Figure 1], [Figure 2], [Figure 3]. Significant nuclear atypia, mitotic activity or necrosis were not seen. The tumor cells showed strong immunoreactivity for alpha smooth muscle actin. ER and PR stains were positive while TTF-1 was negative in the tumor. No other lesions were found elsewhere in the body by appropriate investigations.

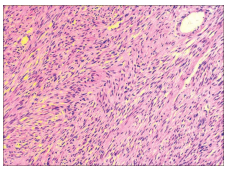

Figure 1 :Fascicles of smooth muscle fibers without atypia or necrosis (H and E, ×100)

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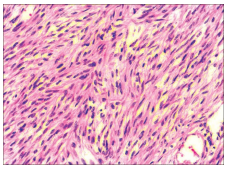

Figure 2 :Smooth muscle fibers with cigar-shaped nuclei and occasional mitosis (H and E, ×200)

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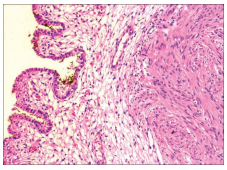

Figure 3 :Sub-pleural lung nodule having spindle cell lesion of similar morphology as seen in uterus (H and E, ×100)

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Discussion

This entity is characterized by multiple small nodules in various sites, mostly lung composed of well-differentiated smooth muscle cells with concomitant or preexisting ureteric leiomyoma. PE Steiner [3] at university of Chicago in 1939 was the first who described this entity namely 'Benign metastasizing fibroleiomyoma as a tumor composed histologically, in both the primary growth and its metastasis, of benign appearing, fully differentiated smooth muscle cells and dense connective tissue.' The term benign metastasizing leiomyoma (BML) was coined by JP Horstman to distinguish the myometrial origin of the disease as cited by Horstman *et al.* [4] There are several hypotheses regarding the origin of the lesion. It could be a benign uterine tumor that spreads through hematogenous route to the lungs or other organs as majority of patients had prior myomectomy or hysterectomy. It could be a low-grade leiomyosarcoma metastasizing to lungs or it could represent primary pulmonary leiomyomatosis unrelated to but coexisting with uterine leiomyomata. Although lung is the most commonly affected organ, BML has also been reported in lymph nodes, deep soft tissues, mesentery, bones, CNS and the heart. [5],[6]

Benign metastasizing leiomyoma is a morphologically benign neoplasm with bland cytological features. Most express estrogen and progesterone receptors with very low Ki-67 index. In our case, the patient presented with an abdominal mass first and pulmonary nodules were discovered soon after hysterectomy. This is in contrast with most of the earlier reported cases in which there was an interval of several years. The pleural nodule, which was resected, had bland cytological features without enhanced mitotic activity or coagulative necrosis. No atypical mitosis was seen. The morphology was similar to that seen in the uterine leiomyoma. The uterine tumors did not reveal any of the criteria of malignancy outlined by the WHO classification system. [7] Hence they were labeled as leiomyomata.

The clinical course of BMT is variable in spite of its, bland cytology. The usual scenario is of indolent disease. However respiratory failure and death may occur as a result of rapid progression. [8] These tumors are responsive to hormonal manipulation, as they express estrogen and progesterone receptors. Luteinizing hormone releasing hormone (LHRH) agonists, anti-estrogens and aromatase inhibitors. Medical treatment including LHRH agonists, anti-estrogens and

aromatase inhibitors have been successful in relieving the symptoms and leading to regression of metastatic lesions. [2], [9] In some cases, extensive tumor debulking and oophorectomy for hormone control have been used. Progesterone treatment has also been shown to be effective in both the prophylaxis against recurrences and regression of leiomyomatous tumors. [10]

In summary, BML has histological and immunohistochemical features of a benign neoplasm. The presence of metastatic nodules is in contrast with a benign nature. However, based solely on the clinical progression this lesion cannot be labeled as sarcoma. In one study, these tumors were proposed to be regarded as borderline tumors of low malignant potential. [11] Most of the patients in the largest series by Kayser *et al,[11]* had median survival of 43 months after lung biopsy and did not die of the disease, even without hormonal therapy. Further detailed studies are required to establish markers, which would identify uterine leiomyomas with risk of development of BML.

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Figures

[Figure 1], [Figure 2], [Figure 3]

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